Pulmonary Artery Aneurysms: Four Case Reports and Literature Review

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Abstract

Aneurysms of the pulmonary artery are proven to be a very rare entity. Association with structural cardiac anomalies, structural vascular anomalies, pulmonary hypertension, vasculitis, and infection has been noted. Surgical intervention of symptomatic aneurysms is recommended. A more detailed study of the natural history of these aneurysms is needed. Here, we report four cases of pulmonary artery aneurysms as well as a brief review of the literature existing on this subject. The first case is of a 41-year-old woman with the aneurysm located 1 cm distal to the pulmonary valve extending to the bifurcation of the main pulmonary artery. The second case is of a 76-year-old woman with a large aneurysm of the main pulmonary artery and the left pulmonary artery. The third case is of a 61-year-old woman with an aneurysm of the common pulmonary artery and right pulmonary artery. The fourth case is of a 28-year-old woman with a 5-cm symptomatic aneurysm extending from the valve up to the pulmonary bifurcation. Surgical excision and reconstruction was ordered for cases 1, 2, and 4.

Keywords
- pulmonary artery
- aneurysm

Aneurysms of the pulmonary arteries (PAs) and trunk are a rare entity. They have been associated with structural cardiac anomalies (especially congenital heart disease), structural vascular anomalies, vasculitis, pulmonary hypertension, and infection, but idiopathic pulmonary artery aneurysms (PAAs) have also been identified. Recently, reports and identification of these clinical entities have increased owing to advances in diagnostic imaging methods such as computed tomography (CT), magnetic resonance imaging (MRI), and echocardiography. However, their natural history has not been extensively studied and remains largely unknown. Here, we present four cases of PAAs (three surgical and one medical) and review the existing literature on the subject.

Case Presentations

Case 1

A 41-year-old woman with a history of diabetes since childhood, hypertension and obesity was noted to have an abnormal electrocardiogram, which led to a cardiac evaluation. She was also noted to have episodes of dyspnea, which were getting more severe. She ultimately had a chest CT, which revealed a dilation of the main PA.

The PA measured approximately 5.6 to 6.5 cm in diameter and the aneurismal zone extended from 1 cm distal to the pulmonary valve to the bifurcation of the main PA. The right and left main PAs appeared normal in caliber. Echocardiography revealed a severely dilated main PA and valve, with mild pulmonary insufficiency and which showed an estimated PA systolic pressure of 43 mm Hg. It was decided that the aneurysm should be removed and repaired surgically. MRI confirmed the findings of the echo and the CT scan.

A standard median sternotomy was performed and the patient was placed on cardiopulmonary bypass. Cross-clamp time was 41 minutes and the lowest core temperature was 28°C. The PAA (►**Fig. 1**) was resected, and the right and left PAs were mobilized. The repair was performed primarily to reconstruct the PA.

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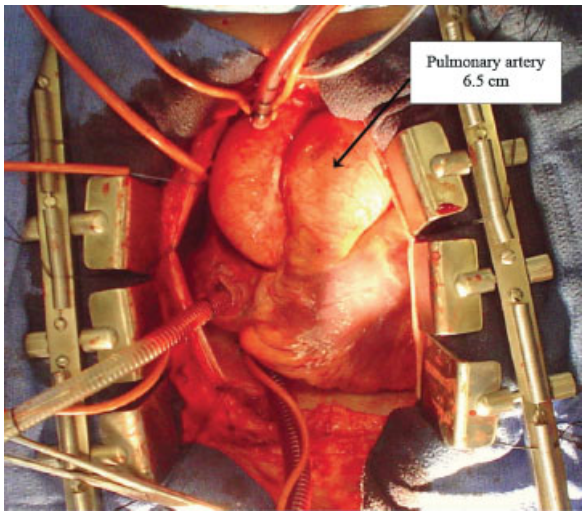

Fig. 1 Intraoperative image of a large pulmonary artery aneurysm.

An exercise (stress) tetrofosmin SPECT imaging, performed 5 years later, showed a normal sized left and right ventricle with normal function and an ejection fraction of 80%.

Case 2

A 76-year-old woman presented with a history of chest discomfort and persistent cough. Past history included hypertension, hypercholesterolemia, hypothyroidism, and thrombophlebitis.

CT and MR angiography revealed a large main PAA (►**Fig. 2**) as well as a large left PA. The main PAA measured 4.4 cm in diameter and the dilatation continued into the left PA, which measured 5.2 cm in diameter. The ascending aorta was mildly dilated at 4.0 cm. The right ventricular wall appeared to be hypertrophied and the right atrium dilated. There was a left atrial clot seen on preoperative echo. Preoperative carotid ultrasound showed a 50% stenosis at the origin of the internal carotid artery. Intraoperative transesophageal echo showed a large patent foramen ovale. It was decided that the aneurysm should be removed and repaired and closure of the atrial septal defect (ASD) performed.

Median sternotomy was performed and the patient was placed on cardiopulmonary bypass. The PAAs were resected by removing a significant circumference of the anterior portions of the arteries by a longitudinal incision down the main PA and onto the left PA. The incision was extended just above the level of the bifurcation, at which point it appeared that the PA narrowed and became normal caliber. The longitudinal incision was closed primarily with running suture. The large ASD was closed primarily with interrupted mattress sutures.

Case 3

A 61-year-old woman came under our care 6 years ago because of a neck bruit. The patient's PA was enlarged, with moderate dilatations of the main pulmonary trunk and right main PA, approximately 4 and 3 cm in greatest dimension, respectively, on contrast-enhanced CT examination (►**Fig. 3**). In addition, calcified coronary arteries, aortic arch calcification, and a "bovine" aortic arch were noted.

It was decided that surgery was not needed, since the pulmonary aneurysm was not massive and was not producing any symptoms. She embarked on serial follow-up.

Case 4

A 28-year-old woman with a medical history of hyperthyroidism, obesity, and childhood asthma presented with unrelenting chest pain. Several weeks previously, she was admitted with pain, which increased with inspiration in her right flank/subscapular region but subsided by hospital day 2 and was discharged on hospital day 4.

CT scan showed a 5-cm aneurysm of the PA. The PA was enlarged from just above the valve up to the pulmonary bifurcation. The dilation was bulbous and extended rightward underneath the aorta and posteriorly. There was also

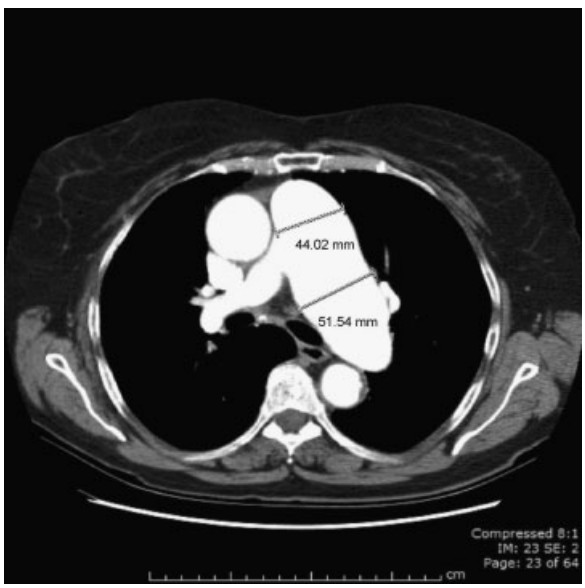

Fig. 2 Computed tomography image showing an aneurysm of the main pulmonary artery and left pulmonary artery. Patient underwent surgical treatment.

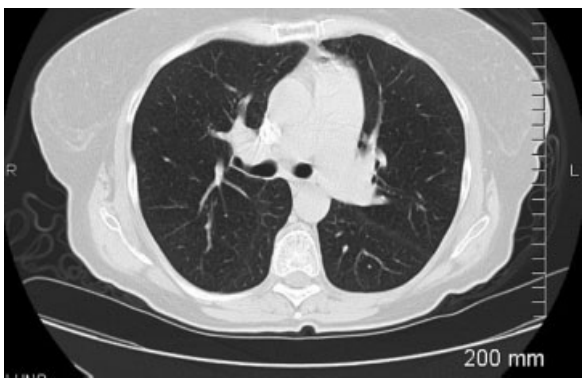

Fig. 3 Computed tomography image showing dilatation of the main, right, and left pulmonary arteries. Patient is currently on medical treatment.

a prominent thrill noted over the PA originating near the pulmonary outflow tract, indicating turbulent flow. Because of the symptomatic nature of her aneurysm and the concern over straining during childbirth, since she had the desire of becoming pregnant, surgical correction was deemed necessary.

Median sternotomy was performed and the procedure was done on cardiopulmonary bypass with myocardial preservation by systemic hypothermia to 32 degrees, topical hypothermia with iced saline, and cold crystalloid cardioplegia given at intervals into the aortic root. Cross-clamp time was 34 minutes and total bypass time was 50 minutes. The aneurysmal segment was excised and the proximal and distal cuffs were fashioned. A primary anastomosis was done with running suture.

After 8 years, the patient is doing very well. She has not had any operation-related problems and has not had any other surgeries since. She is regularly seen in follow-up by her primary care physician.

Discussion

PAAs are a rare entity. In the year 1947, Deterling and Clagett[1] published a review of proximal PAAs over an extended period of 100 years. They reviewed 92,026 autopsy studies and added 17,545 of their own (total of 109,571 cases) and concluded that only 8 cases of PAA had been documented. That translates that the reported incidence was 0.0073%.

Definition

The upper normal limit for the diameter of the main PA on CT is 29 mm and that of the right interlobar artery is 17 mm.[2,3] A PAA that exceeds this dimension can be considered enlarged, and one that exceeds 4 cm can be considered aneurismal (see below).

Classification

PAAs can be classified into proximal (or central) PAAs and peripheral PAAs.

Proximal PAAs involve the pulmonary trunk and the main right and left PAs. Proximal PAAs are defined as a diameter of over 4 cm in the PA trunk.[4] In 66% of PAA patients, pulmonary hypertension is noted.[5] Symptoms are rarely seen unless there are complications such as bronchial or tracheal compression (leading to cough and dyspnea), dissection, or rupture. Also, thrombus can form in the PAA, caused by reduced blood flow velocity. Dissections of the PA are usually associated with pulmonary hypertension. Only 19% of patients with dissection did not have pulmonary hypertension.[6] Eighty percent of dissections occurred in the main pulmonary trunk.[7] The peripheral PAAs encompass the aneurysms located in the intrapulmonary arteries. In a study of 111 cases with PAA, only 22 cases were identified in the peripheral portion making up for 19.8%.[5] Most of the mycotic aneurysms are located in this part, and historically when tuberculosis (TB) was a more prominent disease among the general population, it constituted the most common etiology. The aneurysms caused by TB were named "Rasmussen aneurysms."[8] Peripheral PAA can be considered a life-threatening condition, since its most grave symptom is massive hemoptysis. Even in cases where rupture had not occurred, untreated cases had a very high mortality rate.[9] The treatment modality preferred in these cases is coil embolization, since it is the least invasive therapy especially because surgery poses a very high risk for patients with severe pulmonary hypertension.[10]

Etiology and Pathogenesis

Several underlying conditions assume the role of risk factors and causative agents for developing PAAs. Infection, structural cardiac anomalies, structural vascular anomalies, and pulmonary hypertension are among the most common. Idiopathic, isolated aneurysms also do occur, but they are very rare entity.

Infection

Historically, TB and syphilis played the main role, but with the significant drop in incidence of these diseases nowadays, other organisms have replaced them. In TB, Rasmussen aneurysms are seen in vessels that go through tuberculous cavities; thus, they occur in patients with chronic progressive disease (4 to 5% of chronic TB cases). The mechanism of the aneurysm formation lies in the vessel's wall tissue destruction and replacement by granulomatous tissue, causing thickening and weakening of the arterial wall from the outer to the inner wall.[11] In syphilitic aneurysms, the mechanism of formation involves destruction of the vasa vasorum, which in turn leads to weakening of the vessel wall and appears to induce atherosclerotic changes.[12] TB and syphilis are largely of historical significance in the present era as causes of PAA. There is also a strong link between mycotic aneurysms and pulmonary hypertension. In a vessel wall already under stress, septic emboli can lodge and cause aneurysmal formation.[13]

Structural Cardiac Anomalies

Most PAAs are congenital (the most common cause of PAAs), but acquired cases have also been reported. According to the two major proximal PAA autopsy studies, congenital cardiac abnormalities were found in 56% of the cases.[1] The most common defects were, in order of highest occurrence, patent ductus arteriosus (PDA), ASD, and ventricular septal defect. Other less common congenital anomalies include tetralogy of Fallot, transposition of the great vessels, and bicuspid aortic valve. In the case of a PDA, there is a left-to-right shunt observed that causes a "jet stream" to hit the wall of the PA, bringing about local injury and weakness of the arterial wall.[1] Acquired rather than congenital-associated lesions include mitral stenosis, pulmonic stenosis, tricuspid insufficiency, and pulmonic insufficiency; these are reflective of conditions of excess volume or pressure load on the pulmonary thought to play a role in the dilatation leading to aneurysm formation.

Structural Vascular Abnormalities

In comparison with structural cardiac abnormalities, which most often are congenital, the majority of structural

vascular abnormalities leading to PAA are acquired degenerative diseases. Medionecrosis (cystic medial necrosis) and atherosclerosis are the most prominent in the literature. Their pathogenesis is the result of a continual process of damage to and repair of the vascular structures.[14] Medionecrosis or atherosclerosis has been identified in the majority of the cases of dissecting PAA reported to date. To add, medionecrosis of the PA differs from the aorta in the fact that there is no male sex predisposition, but otherwise it is very similar to the aortic variant.[15,16] Marfan syndrome (MFS) also plays a significant role in structural vascular abnormalities leading to PAA; although MFS mainly involves the aorta, PA cases have also been identified.[17] Last but not the least, vasculitis seems to be an obvious precursor of PA aneurysms. Behçet disease and giant cell arteritis are the leading vasculitic variants causing PAA.[18,19] It has been noted that aneurysms due to Behçet disease can spontaneously regress following medical treatment of the disease. Surgery is discouraged in Behçet disease due to recurrent false aneurysm formation at the anastomotic sites.[20] Another rare vasculitic disease is Hughes–Stovin syndrome, which includes recurrent thrombophlebitis and PAA formation and rupture.[21]

Pulmonary Hypertension

Pulmonary hypertension can be classified into precapillary (idiopathic, schistosomiasis), capillary (chronic obstructive pulmonary disease, interstitial lung disease, fibrothorax), and postcapillary (mitral valve stenosis, left heart failure, left atrial myxoma, veno-occlusive disease). Idiopathic primary pulmonary hypertension is a rare disease and is characterized by plexiform lesions, endothelial cell proliferation, and concentric laminar intimal fibrosis of the PA.[22] Another important risk factor for pulmonary hypertension is chronic pulmonary embolism. Pulmonary hypertension predisposes to PAA in some patients.

Trauma

Trauma can be divided into extravascular and endovascular. Both blunt and penetrating trauma account for the extravascular causes of PAA, penetrating stab wounds being the most frequent.[23] Endovascular trauma is mainly iatrogenically induced. Malpositioned Swan-Ganz catheters are the most common cause. This complication occurs when the catheter has been inserted too far into a pulmonary arterial branch. The pathogenesis is erosion of the tip of the catheter into the wall of the artery, causing weakening and dilatation. In a prospective study on 500 patients, it was proven that the incidence of rupture and hemorrhage after the Swan-Ganz catheter is 0.2%.[24] Other iatrogenic causes include chest tube insertion, conventional angiography, and surgical resection or biopsy.

Clinical Symptoms and Diagnosis

The clinical symptoms vary and are related to the underlying etiologies, location, and size of these PAAs. Hemoptysis is the most frequently identified symptom resulting from rupture, and it is mostly fatal. Dyspnea and cough are observed when the aneurysm compresses the trachea or the bronchi. Fever is seen mainly in mycotic aneurysms. Sometimes, a harsh systolic murmur can be heard over the left second and third intercostal space from pulmonic valve disease.[25] As already noted, the most catastrophic outcome of PAA of the main PA is aneurysmal rupture or dissection. Laplace law dictates that wall stress, which constitutes the most important factor for progression to rupture, is directly proportional to the pressure and radius of a vessel wall and is inversely proportional to the wall thickness.[26] In the absence of significant pulmonary regurgitation or stenosis, which can cause right ventricular dysfunction, pulmonary hypertension, or a left-to-right shunt, the risk of aneurysmal rupture seems to be very small.[27,28]

Diagnosis

The gold standard for the diagnosis of PAAs has been pulmonary angiography. Angiography is invasive and identifies only the interior of the aneurysm, the segment that has active flow. With recent technical advances, other modalities have largely supplanted angiography for diagnosis of PAA. Spiral CT is an excellent diagnostic modality, as it can demonstrate the patent lumen as well as any mural thrombus or other abnormalities of the vessel wall. Spiral CT also has the ability of multiplanar reconstruction that can provide very useful information to the surgeon for planning surgery. For optimal imaging in the workup, echocardiography and MRI should be included.[29]

Treatment

Treatment can be either conservative (medical) or surgical.

Surgical repair is recommended if the aneurysms are large, $>$ 6 cm, or if they are symptomatic, regardless of the size, because the risk of rupture or dissection is high in the case of symptoms.[30] These criteria for intervention are based on limited natural history data. For proximal PAAs coexisting with pulmonary hypertension, the interventional technique can be the treatment of the pulmonary hypertension alone. Although medical treatment alone in complicated cases can pose inadequate.[31]

The surgical techniques that have been described include aneurysmorrhaphy or arterioplasty, pericardial patch reconstruction, and interposition grafting with allografts or synthetic textile grafts as treatment methods for aneurysms of the main PA.[32,33] Recently, steel coil embolization has shown promising results for treatment of peripheral PAAs.[34] Peripheral PAAs in the past were treated with lobectomy or aneurysmectomy, but currently endovascular coil embolization is preferred, since it is less invasive and fewer complications have been observed.[35]

Conservative treatment is recommended for patients who do not experience symptoms of PAAs and have aneurysms less than 6 cm in diameter. Patients diagnosed with pulmonary hypertension should be treated medically to lower the pulmonary pressure. As mentioned earlier, aneurysms caused by Behçet disease seem to regress after anti-inflammatory treatment for this disease.

Conclusions

PAAs are rare. Congenital heart disease and pulmonary hypertension are the leading causes in the present era. Rupture and PA dissection can occur. Surgical intervention is recommended for large aneurysms of the main PA trunk/main PAs or for large aneurysms in those locations. Better clarification of the natural history of these rare arterial aneurysms is needed.

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